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외음부의 혈관근섬유아세포종 1예

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A case of Angiomyofibroblastoma

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Abstract : Angiomyofibroblastoma of the vulva is a rare mesenchymal tumor. It is characterized by superficial and slow growth, low propensity for local recurrence and often misdiagnosed as a Bartholin's gland cyst and aggressive angiomyxoma. We report a case of angiomyofibroblastoma of the vulva in a 23-year-old female patient. She presented with two lobulating left vulvar masses for about 2 years. Initially the mass was small and reminiscent of a Bartholin's cyst but grew rapidly during the recent two months. Grossly the tumor consisted of large two lobulating masses with superficial ulceration.

Microscopically there were alternating hypercellular and hypocellular edematous zones, in which abundant capillary blood vessels were noted. Immunohistochemically the stromal cells showed positive reaction for vimentin, desmin, and smooth muscle actin.

Key Words : Angiomyofibroblastoma, Vulva

Angiomyofibroblastoma of the genital region is a relatively recently described tumor of the superficial soft tissues with a marked preference for female patients.[1] Since symptoms of benign and malignant conditions in the vulvar area are similar, early diagnosis and treatment is important.

This report presents several interesting features of an angiomyofibroblastoma of the vulva with respect to the origin of the tumor and histologic and immunohistochemical findings.

Case report

A 23-year-old woman having a vulvar mass over a period of two years visited our clinic in February 2001. The mass was initially confused with a Bartholin's cyst but grew rapidly during the recent one month to have about 7 to 8 times the size of that at the intial diagnosis. The genital examination showed no other abnormal findings and the tumor located subcutaneously in her left labial region consisting of two lobulated masses. It was well defined, soft to cystic in consistency, mobile, nontender, and superficially ulcerated (Fig. 1). The patient had no remarkable previous medical record and no history of medication. General physical and abdominal examination revealed no abnormalities.

책임저자 : 고민환

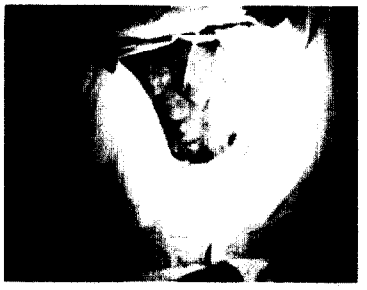

Fig. 1. Preoperative feature of the mass. Note the two lobulating masses with superficial ulceration.

Fig. 2. The tumor was removed by local excision.

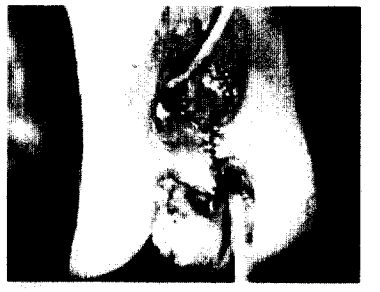

Fig. 3. The remaining tissue was trimmed and the surgical wound was closed with a drainage insertion.

A vaginal examination showed normal findings without an adnexal mass. There was no inguinal lymphadenopathy. We did not check pelvic CT or MRI before removal of the mass.

We performed a simple excision of the mass (Fig. 2). The whole vulvar mass was excised and silastic drain was applied (Fig. 3).

Postoperative course of the patient was uncomplicated and there is currently no evidence of recurrence one year after surgery.

The tumor measured 16.0×15.5×7.0cm in size and 425.0 g in weight. The cut surface showed gray-to-pinkish solid appearance with variable sized cystic spaces and also showed myxoid appearance simulating leiomyoma with myxoid degeneration (Fig. 4).

Fig. 4. Grossly the tumor was well circumscribed and showed solid, myxoid appearance.
The tumor measured 16 × 15.5 × 7.0cm in size.

On microscopic examination the tumor showed alternating hypercellular and hypocellular edematous areas containing small vessels. The tumor cells were spindle-to-oval cells in shape with bland nuclei and showed a tendency to perivascular aggregation (Fig. 5, 6). The cells were immunoreactive to vimentin, desmin, and smooth muscle actin (Fig. 7).

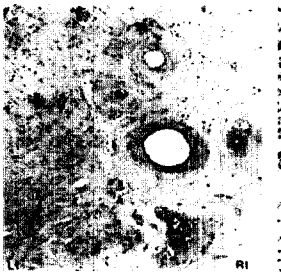

Fig. 5. Histologically the tumor showed alternating hypercellular (left) and hypocellular (right) areas with perivascular accentuation of stromal cells(H-E stain, ×100).

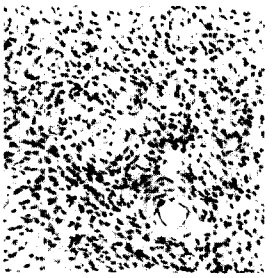

Fig. 6. The tumor showed a focal area of compact proliferation of bland-looking stromal cells(H-E stain, × 200).

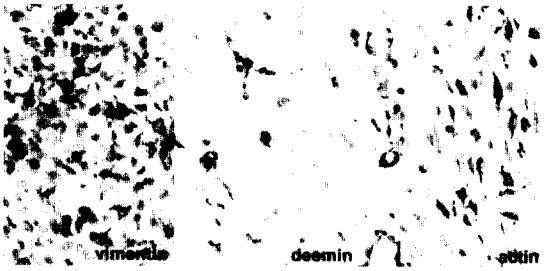

Fig. 7. Immunohistochemical staining revealed positive reaction for vimentin, desmin, and actin(×400).

Discussion

Angiomyofibroblastoma is a rare, recently

described, soft tissue tumor that occurs mainly, but not exclusively, in the vulvar area.[2] As in our case, patients are usually premenopausal and present with a vulvar mass initially misdiagnosed as a Bartholin's gland cyst. Angiomyofibroblastoma should be differentiated from other neoplasms of the vulva in which radical surgical treatment is indicated. Local excision of the tumor is the treatment of choice.[4] Except for one case with malignant transformation of angiomyofibroblastoma in an 80-year-old woman, there are currently no published reports of local recurrence or metastatic disease of angiomyofibroblastoma in the literature.[4]

The most important differential diagnosis of angiomyofibroblastoma is aggressive angiomyxoma (Table 1). Angiomyofibroblastoma is distinguished

Table 1. Comparison of angiomyofibroblastoma with aggressive angiomyxoma[2]

	Angiomyofibroblastoma	Aggressive angiomyxoma
Clinical features		
Age	25-54 yrs(mean 36.3 yrs)	18-63 yrs(mean 32.1 yrs)
Presentation	Vulvar mass	Vulvar/pelvic mass
Duration of symptoms	10 weeks - 8 years	Usually a few months
Location of lesion	Vulva	Vulva, vagina, perineum or pelvic soft tissues
size of lesion	0.5-12cm (usually < 5 cm)	3-60cm (usually > 5ßØ)
Behavior	No recurrence after simple excision	Local recurrence in >70% of cases, often within 2 years (but sometimes delayed)
Pathology		
Borders	Well circumscribed	Infiltrative; at most partly circumscribed
Blood vessels	More numerous: mostly capillaries, with some thin-walled ectatic/cavernous veins	Small to medium-sized vessels, many of which are thick-walled or hyalinized
Stromal cells	More abundant and with perivascular accentuation; wavy spindle, plump spindle, oval, and multinucleated giant cells	Low cellularity; stellate or spindled stromal cells, with delicate cytoplasmic processes
Stroma	Edematous to collagenous; extravasation of erythrocytes uncommon	Myxoid to collagenous, often with extravasation of erythrocytes
Immunostaining		
Vimentin	Positive	Positive
MSA/ASMA	Negative	Variable; usually negative
Desmin	Positive	Negative

from aggressive angiomyxoma by its well circumscribed border and higher cellularity, by the frequent presence of plump stromal cells, and by a lesser degree of stromal myxoid change.[8] In contrast to angiomyofibroblastoma aggressive angiomyxoma is a deeply seated tumor with an infiltrative growth pattern which frequently results in entrapment of mucosal glands and nerves.[6] In the present case immunohistochemical staining showed the tumor cells were positive for desmin, vimentin, and smooth muscle actin.

The size of the angiomyofibroblastoma in the vulvar area varies from 0.5cm to 12cm, however in our case, the tumor measured 16cm in its greatest dimension and thought to be the largest tumor ever reported.[9]

Angiomyofibroblastoma is relatively a newly recognized tumor on the expanding spectrum of benign soft tissue tumors and should be considered in the differential diagnosis of vulvar lesions in elderly postmenopausal patients. Differentiation of the tumor from other diseases such as aggressive angiomyxoma, where more aggressive surgical approaches are required, is mandatory.[9]

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■ 국문 초록 ■

외음부의 혈관근섬유아세포종(angiomyofibroblastoma)는 비교적 드문 중간엽성 종양이다. 혈관근섬유아세포종은 표재성이며 느린 성장을 보이고 국소 재발이 적은 경향이 있지만 종종 바르톨린샘 낭종(Bartholin's gland cyst)과 침윤성 혈관점액종(aggressive angiomyxoma)으로 오진하는 경우가 있어 감별을 요한다. 최근 저자들은 23세 여성의 외음부에서 발생한 혈관근섬유아세포종을 경험하였는데 환자는 약 2년간 두 개의 분엽을 가진 왼쪽 외음부 종괴를 호소하였다. 처음에는 바르톨린샘 낭종으로 생각하였으나 최근 2개월간 빠르게 성장하였으며 육안적으로 보았을 때 종양은 표재성 궤양을 동반한 큰 두 개의 분엽을 이루고 있었다.

광학 현미경 소견상 종양내에는 풍부한 모세혈관과 함께 주위 세포밀도가 다르게 관찰되는 부위가 있었다. 면역조직화학적 염색에서 종양의 기질세포는 vimentin, desmin, smooth muscle actin에 양성을 보였다.

핵심단어 : 혈관근섬유아세포종, 외음부